LYME DISEASE

DISEASE REPORTING

In Washington

DOH receives 7 to 18 reports of Lyme disease per year.

Almost all Washington cases are the result of outdoor exposure in counties west of the Cascade Mountains in the Cascade foothills, or out of state, reflecting the distribution of the *Ixodes* tick vector and its deer and rodent reservoirs. The rate of infection in Washington ticks is low, but high in Atlantic states, Minnesota and Wisconsin.

The tick must remain attached for at least 24 hours to transmit infection.

Purpose of reporting and surveillance

- To educate people about how to reduce their risk of infection.
- To better characterize the epidemiology of this disease.
- To identify endemic geographic areas in Washington.

Reporting requirements

- Health care providers: notifiable to Local Health Jurisdiction within 3 work days
- Hospitals: notifiable to Local Health Jurisdiction within 3 work days
- Laboratories: no requirements for notification
- Local health jurisdictions: notifiable to DOH Communicable Disease Epidemiology within 7 days of case investigation completion or summary information required within 21 days

CASE DEFINITION FOR SURVEILLANCE

Clinical criteria for diagnosis

A systemic, tickborne disease with protean manifestations, including dermatologic, rheumatologic, neurologic, and cardiac abnormalities. The best clinical marker for the disease is the initial skin lesion (i.e., erythema migrans [EM]) that occurs in 60%–80% of patients.

Laboratory criteria for diagnosis

- Isolation of *Borrelia burgdorferi* from a clinical specimen or
- Demonstration of diagnostic immunoglobulin M or immunoglobulin G antibodies to *B. burgdorferi* in serum or cerebrospinal fluid (CSF). A two-test approach using a

sensitive enzyme immunoassay or immunofluorescence antibody followed by Western blot is recommended.

Case definition

- Confirmed: a) a case with EM or b) a case with at least one late manifestation (as defined below) that is laboratory confirmed.
  This surveillance case definition was developed for national reporting of Lyme disease; it is not intended to be used in clinical diagnosis. Definition of terms used in the clinical description and case definition:
  - *Erythema migrans. For purposes of surveillance, EM is defined as a skin lesion that typically begins as a red macule or papule and expands over a period of days to weeks to form a large round lesion, often with partial central clearing. A single primary lesion must reach ≥5 cm in size. Secondary lesions also may occur. Annular erythematous lesions occurring within several hours of a tick bite represent hypersensitivity reactions and do not qualify as EM. For most patients, the expanding EM lesion is accompanied by other acute symptoms, particularly fatigue, fever, headache, mildly stiff neck, arthralgia, or myalgia. These symptoms are typically intermittent. The diagnosis of EM must be made by a physician. Laboratory confirmation is recommended for persons with no known exposure.*
  - *Late manifestations. Late manifestations include any of the following when an alternate explanation is not found:*
    - *Musculoskeletal system. Recurrent, brief attacks (weeks or months) of objective joint swelling in one or a few joints, sometimes followed by chronic arthritis in one or a few joints. Manifestations not considered as criteria for diagnosis include chronic progressive arthritis not preceded by brief attacks and chronic symmetrical polyarthritis. Additionally, arthralgia, myalgia, or fibromyalgia syndromes alone are not criteria for musculoskeletal involvement.*
    - *Nervous system. Any of the following, alone or in combination: lymphocytic meningitis; cranial neuritis, particularly facial palsy (may be bilateral); radiculoneuropathy; or, rarely, encephalomyelitis. Encephalomyelitis must be confirmed by demonstration of antibody production against* B. burgdorferi *in the CSF, evidenced by a higher titer of antibody in CSF than in serum. Headache, fatigue, paresthesia, or mildly stiff neck alone are not criteria for neurologic involvement.*
    - *Cardiovascular system. Acute onset of high-grade (2° or 3°) atrioventricular conduction defects that resolve in days to weeks and are sometimes associated with myocarditis. Palpitations, bradycardia, bundle branch block, or myocarditis alone are not criteria for cardiovascular involvement.*
  - *Exposure. Exposure is defined as having been (≤30 days before onset of EM) in wooded, brushy, or grassy areas (i.e., potential tick habitats) in a county in which Lyme disease is endemic. A history of tick bite is not required.*
  - *Disease endemic to county. A county in which Lyme disease is endemic is one in which at least two confirmed cases have been previously acquired or in which established populations of a known tick vector are infected with* B. burgdorferi.

A. DESCRIPTION

1. Identification

This tickborne, spirochetal, zoonotic disease is characterized by a distinctive skin lesion, systemic symptoms and neurologic, rheumatologic and cardiac involvement that occur in varying combinations over a period of months to years. The early symptoms are intermittent and changing. The illness typically begins in the summer, and the first

manifestation in about 90% of patients appears as a red macule or papule that expands slowly in an annular manner, often with central clearing. This distinctive skin lesion is called erythema migrans (EM; formerly erythema chronicum migrans). EM may be single or multiple. To be considered significant for case surveillance purposes, the EM lesion must reach 5 cm in diameter. With or without EM, early systemic manifestations may include malaise, fatigue, fever, headache, stiff neck, myalgia, migratory arthralgias and/or lymphadenopathy, all of which may last several weeks or more in untreated patients.

Within weeks to months after onset of the EM lesion, neurologic abnormalities such as aseptic meningitis and cranial neuritis-including facial palsy, chorea, cerebellar ataxia, motor or sensory radiculoneuritis, myelitis and encephalitis-may develop; symptoms fluctuate, may last for months and may become chronic. Cardiac abnormalities (including atrioventricular block and rarely, acute myopericarditis or cardiomegaly) may occur within a few weeks after onset of EM. Weeks to years after onset (mean, 6 months), intermittent episodes of swelling and pain in large joints, especially the knees, may develop and recur for several years; chronic arthritis may occasionally result. Similarly, sometimes following long periods of latent infection, chronic neurologic manifestations may develop and include encephalopathy, polyneuropathy or leukoencephalitis; the CSF often shows lymphocytic pleocytosis and elevated protein levels, while the electromyogram is usually abnormal.

Diagnosis is currently based on clinical findings supported by serologic tests performed in two stages, by IFA, ELISA, then Western immunoblot. Serologic tests, which are poorly standardized, must be interpreted with caution. They are insensitive during the first several weeks of infection and may remain negative in people treated early with antibiotics. An ELISA for IgM antibodies that uses a recombinant outer surface protein C (rOspC) has been shown to be more sensitive for early diagnosis than either whole cell ELISA or immunoblot assay. Test sensitivity increases when patients progress to later stages of the disease, but a small proportion of chronic Lyme disease patients may remain seronegative. Cross-reacting IFA and ELISA antibodies may cause false positive reactions in patients with syphilis, relapsing fever, leptospirosis, HIV infection, Rocky Mountain spotted fever, infectious mononucleosis, lupus or rheumatoid arthritis. The specificity of serologic testing is enhanced by immunoblot testing of all specimens that are positive or equivocal on IFA or ELISA. The etiologic agent, *Borrelia burgdorferi*, grows at 33°C (91.4°F) in the Barbour, Stoenner, Kelly (BSK) medium; other species causing Lyme-like disease may not grow well in this medium. Isolation from blood and tissue biopsies is difficult, but biopsies of the EM lesions may yield the organism in 80% or more of cases. By PCR, *B. burgdorferi* genetic material has been detected in synovial fluid, CSF, skin and other tissues, blood and urine; however, the usefulness of PCR in the routine management of Lyme disease patients has yet to be verified.

2. Infectious Agent

The causative spirochete of North American Lyme disease, *B. burgdorferi*, was identified in 1982. Three genomic groups of *B. burgdorferi* sensu lato have now been identified in Europe; they have been named *B. burgdorferi* sensu stricto, *B. garinii* and *B. afzelii*.

3. Worldwide Occurrence

In the US, endemic foci exist along the Atlantic coast and are concentrated between Massachusetts and Maryland; in the upper midwest, an expanding focus is currently concentrated in Wisconsin and Minnesota; and in the west, in some areas of California and Oregon. Currently, increasing recognition of the disease is redefining endemic areas; cases have been reported from 47 states and from Ontario and British Columbia, Canada. Elsewhere, it has been found in Europe, the former Soviet Union, China and Japan.

Initial infection occurs primarily during summer, with a peak in June and July, but may occur throughout the year, depending on the seasonal abundance of the tick in different geographic areas. The distribution of the majority of cases coincides with the distribution of *Ixodes scapularis* (formerly called *I. dammini*) ticks in the eastern and midwestern US, *I. pacificus* in western US, *I. ricinus* in Europe and *I. persulcatus* in Asia. Dogs, cattle and horses develop systemic disease that may include the articular and cardiac manifestations seen in human patients. The explosive repopulation of white-tailed deer in the eastern US by white-tailed deer has been linked to the spread of Lyme disease in this region.

4. Reservoir

Certain ixodid ticks through transstadial transmission. Wild rodents, especially Peromyscus spp. in the northeastern and midwestern US and *Neotoma* spp. in the western US maintain the enzootic transmission cycle. Deer serve as important maintenance mammalian hosts for vector tick species. Larval and nymphal ticks feed on small mammals, and adult ticks feed primarily on deer. The majority of Lyme disease cases result from bites by infected nymphs.

5. Mode of Transmission

Tickborne; in experimental animals, transmission by *I. scapularis* and *I. pacificus* usually does not occur until the tick has been attached for 24 hours or more; this may also be true in humans.

6. Incubation period

For EM, from 3 to 32 days (mean 7 to 10 days) after tick exposure; however, the early stages of the illness may be inapparent, and the patient may present with later manifestations.

7. Period of communicability

No evidence of natural transmission from person to person. There are rare case reports of congenital transmission, but epidemiologic studies have not shown a link between maternal Lyme disease and adverse outcomes of pregnancy.

8. Susceptibility and resistance

All persons are probably susceptible. Reinfection has occurred in those treated with antibiotics for early disease.

B. METHODS OF CONTROL

1. Preventive measures:

a. Educate the public about the mode of tick transmission and the means for personal protection.
b. Avoid tick infested areas when feasible. To minimize exposure, wear light colored clothing that covers legs and arms so that ticks may be more easily seen; tuck pants into socks and apply tick repellent such as diethyltoluamide (Deet, Autan) to the skin or permethrin (a repellent and contact acaricide) to pant legs and sleeves.
c. If working or playing in an infested area, search the total body area daily, do not neglect haired areas, and remove ticks promptly; these ticks may be very small. Remove any attached ticks by using gentle, steady traction with forceps (tweezers) applied close to the skin to avoid leaving mouth parts in the skin; protect hands with gloves, cloth or tissue when removing ticks from humans or animals. Following removal, cleanse the attachment site with soap and water.
d. Measures designed to reduce tick populations on residential properties are available (host management, habitat modification, chemical control), but are generally impractical on a large-scale basis.
e. During the late 1990s, two Lyme disease vaccines were developed that use recombinant *B. burgdorferi* lipidated outer-surface protein A (rOspA) as immunogen. As of late 1999, one of these vaccines was licensed by the FDA for persons aged 15-70 years in the US. This vaccine is administered on a 3 dose schedule of 0, 1, and 12 months and was found to be safe (does not cause chronic arthritis) and is 76% effective in preventing overt Lyme disease after three doses. Information regarding vaccine safety and efficacy beyond the transmission season immediately after the third dose is not available. Thus, as of late 1999, the duration of protective immunity and need for booster doses beyond the third dose was unknown. [*Note: as of February 25, 2002 Glaxo, the only US manufacturer of Lyme disease vaccine discontinued production of its vaccine. These recommendations apply if any vaccine is currently in use.]
    i. Vaccine induced anti-rOspA antibodies routinely cause false positive ELISA results for Lyme disease. However, experienced laboratory workers, through careful interpretation of the results of Western blot assay, can usually discriminate between *B. burgdorferi* infection and previous rOspA immunization, because anti-OspA antibodies do not develop after natural infection.
    ii. Lyme disease vaccine does not protect all recipients against infection with *B. burgdorferi* and offers no protection against other tickborne diseases. Decisions regarding the use of vaccine should be based on individual

assessment of the risk of exposure to infected ticks and on careful consideration of the relative risks and benefits of the vaccine compared with other protective measures that include early diagnosis and treatment of Lyme disease.

iii. Risk assessment should include consideration of the geographic distribution of Lyme disease. The areas of highest risk in the US are concentrated within some northeastern and north central states. However, the risk for Lyme disease differs not only between regions, states and counties within states, but even within counties and townships. Detailed information about the distribution of Lyme disease risk within specific areas is best obtained from state and local public health authorities.

iv. In areas of moderate to high risk, immunization should be considered for persons aged 15-70 years who engage in activities (e.g., recreational, property maintenance, occupational or leisure) that result in frequent or prolonged exposure to tick infested habitats. Lyme disease vaccine may be considered for persons aged 15-70 years who are exposed to tick infested habitats but whose exposure is neither frequent nor prolonged. The benefit of Lyme disease vaccine beyond that provided by basic personal protection and early diagnosis and treatment of infection is uncertain. Lyme disease vaccine is not recommended for persons who have minimal or no exposure to tick infested habitats.

2. Control of patient, contacts and the immediate environment:

a. Report to local health authority.
b. Isolation: None.
c. Concurrent disinfection: Carefully remove all ticks from patients.
d. Quarantine: None.
e. Immunization of contacts: None applicable.
f. Investigation of contacts and source of infection: Studies to determine source of infection are indicated when cases occur outside a recognized endemic focus.
g. Specific treatment: For adults, the EM stage can usually be treated effectively with doxycycline (100 mg twice daily) or amoxicillin (500 mg 3-4 times daily). For localized EM, 2 weeks of therapy is usually sufficient; for early disseminated infection, 3-4 weeks of therapy should be given. Children less than 9 years of age can be treated with amoxicillin, 50 mg/kg/day in divided doses, for the same period of time as adults. Cefuroxime axetil or erythromycin can be used in those who are allergic to penicillin or who cannot take tetracyclines. Lyme arthritis can usually be treated successfully with a 4-week course of the oral agents. However, objective neurologic abnormalities, with the possible exception of facial palsy alone, are best treated with IV ceftriaxone, 2 g once daily, or IV penicillin, 20 m.u. in 6 divided doses, for 3-4 weeks. Treatment failures may occasionally occur with any of these regimens and retreatment may be necessary.

3. Epidemic measures

In hyperendemic areas, particular attention should be paid to identification of the tick species involved and the areas infested, and to recommendations in B1a through B1c, above.

4. International measures

WHO Collaborating Centres.